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Case report

Intraabdominal Desmoid Tumor: A Case Report

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ABSTRACT

Desmoid tumors (DTs) represent approximately 0.03% of all tumors and 3% of soft tissuetumors. DTsare benign neoplasms of fibroblastic origin and characterized by lack of a capsule. They are nonmetastatic and locally aggressive.However, because of this local infiltration and compression of surrounding structures, a high recurrence rate exists and in anatomic locations with restricted access to surgical resection DT scan lead to death. Herewith we are presenting a rare case of desmoid tumor in a middle aged woman who presented with lower abdominal pain and swelling of about 04-05 months. Past history revealed recurrent surgical interventions for the same for about 2 two times. The tumor with attached fibromuscular tissue was irregular, gray white, with no signs of a capsule. Histopathological examination of the tumor showed features of desmoid tumor. Follow up for a prolonged period has to be done to know it's recurrence.

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1. Introduction

Desmoid tumors (DTs)are rare, benign, soft-tissue tumors characterized by fibroblastic proliferation within a collagen matrix[1].They are characterized by highly aggressive behavior and local invasiveness[2]. DTs are histologically benign, non-encapsulated tumors consisting of well-differentiated fibroblastic cells in a collagen matrix ; they arise from the surface of a fascial or musculo-aponeurotic structure[3]. Despite their invasive potential and high rate of local recurrence, wide surgical excision remains the management of choicesince the results of radiotherapy and chemotherapy have been either disappointing or uncertain [4].

2.Case Report

A thirty year old female came with lower abdominal swelling and vague pain with steady growth over six month period. She was operated for similar abdominal swelling twice. Family history was non-contributory. There was no palpable lymphadenopathy. The patient had noted no signs of obstruction or problems with intestinal transit. Hematological and biochemical parameters were with in normal limits. Ultrasound showed the tumor to be heterogeneous mass. FNAC findings were suggestive of benign spindle cell tumor. Surgery was performed with suspicion of Desmoid tumor.

Gross examination of the excised tissue showed an irregular, dense gray white tumor with attached fibromuscular tissue measuring 7 x5 x4 cms. There were no signs of a capsule and no evidence of necrosis.Cut section of the tumor showed homogenous gray white appearance. There were no enlarged nodes surrounding the tumor. The transection margin was tumor-free.Histologic sections showed proliferation of spindle-shaped or stellate cells, with a fasciculate and storiform growth pattern within a background of myxoid intercellular matrix(Fig.No-1). At the periphery of tumor area, spindle cells were entrapped by skeletal muscle fibers in irregular and inter digitating fashion (Fig.No-2). Individual cells were showing features of well-differentiated fibroblastic cellsin a collagen and myxoid matrix (Fig.No-3). Glassy, hyalinized, and keloid-like collagen fibers were also present focally. There was no evidence of atypical mitosis, necrosis and hemorrhage. Finally a diagnosis of inttraabdominal desmoid tumor was made.

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3.Discussion

Desmoid tumor also known as aggressive fibromatosis (AF) is a rare neoplasm arising from deep musculo-aponeurotic structures[5]. This slow-growing fibrous tumor may arise at any site in the body. It affects 2 to 4 per 100000 persons annually and may occur sporadically or be associated with familial adenomatous polyposis in Gardner syndrome. In the general population, the incidence of desmoids is about 3 per million per year, and the tumors are mainly located in the extremities or in the abdominal wall. Of all patients presenting with a desmoid tumor, at least 7.5% has FAP or will develop FAP later in life. Desmoids range from small, indolent, or even regressive tumors to large and progressive growing neoplasms causing obstruction of vital organs. DTs do not metastasize, although they can present as multifocal disease[6].

Affected patients mostly fall within the age range 10-40 years. Trauma may have a triggering effect in the development of the tumors and the tumors may be solitary or multiple. The clinical behavior and natural history of desmoid tumors remain unpredictable and enigmatic: while in some patients it progresses rapidly and aggressively, in others it is more indolent and may remain stable without any subsequent problem for some time. Most DTsare aggressively invade surrounding tissues and organs or may compress surrounding structures. DTsoften arise from the rectus abdominis muscle in postpartum women and in scars of previous abdominal incisions. Every patient with desmoid tumor should be evaluated for the presence of associated polyposis syndrome by taking a detailed family history, performing colonoscopy and possibly upper Gastrointestinal (GI) endoscopy. The patient in this report did not have a family history of polyposis [3],[6].

DTshave been classified into three main subsets, according to their location: extra-abdominal (60% of cases), abdominal wall (25%), and intra-abdominal (15%) tumors[5].The generic term fibromatosis was originally proposed by Stout[7] for a group of related conditions having in common the following features

1. Proliferation of well-differentiated fibroblasts
2. Infiltrative growth pattern
3. Presence of variable amount of collagen between the proliferating cells
4. Lack of cytologic features of malignancy
5. Scanty or absent mitotic activity
6. Aggressive clinical behavior characterized by repeated local recurrence but lack of capacity to metastasize distantly [7], [8].

Grossly these lesions are often large, firm, and whitish with ill-defined outlines and an irregularly whorled cut surface. Microscopically, most of the proliferating cells have features intermediate between those of fibroblasts and smooth muscle cells [8],[9] (i.e., of myofibroblasts). This myofibroblastic nature is responsible for contracture clinically.Ultrastructural study of fibromatosisdescribed intracytoplasmic collagen formation.Clonal chromosomal aberrations are present in approximately half of the deep-seated fibromatosis but only in 10% of those located superficially. Trisomies 8 and 20 and loss of 5q material represent recurring changes. Another molecular difference between superficial and deep fibromatosis is the lack of beta-catenin and APC gene mutations [8]. Other light microscopic features commonly encountered in DTs are a perivascular lymphocytic infiltrate located at the advancing edge of the lesion and thick-walled vessels sharply outlined from the surrounding tissue. Dystrophic calcification and metastatic ossification have also been described [8].

Deep-seated fibromatosis are potentially aggressive. There is little correlation between the cellularity or other microscopic features of these lesions and their biologic behavior. Well differentiated fibrosarcoma can be differentiated from DTsby the presence of atypical cytologic features and / or a significant number of mitotic figures (more than one per high power field) [8]. Most of the DTsare in intimate contact with skeletal muscles-hence their designation as musculo-aponeurotic fibromatosis [10].

Management of patients with DTs is difficult and many issues remain controversial, mainly regarding early detection, the role, type and timing of surgery, and the value of non-operative therapies. Women have been found to be more likely to require multiple desmoid tumor resections than men, an observation which supports the hypothesis that estrogens stimulate desmoid growth [3]. The treatment of choice is a prompt radical excision, including a wide margin of involved tissue. The incidence of local recurrence is lower in fibromatosis of the abdominal wall than in those located elsewhere [11]. A higher incidence of recurrence was seen in young individuals and in those patients with large size tumors[8],[11]. Radiation therapy may be effective in achieving local control [8].

4.Conclusion:

Desmoid tumors are rare in clinical practice and their management remains quite challenging due to their variable clinical behavior. This case highlights the importance of careful history taking, lateral thinking, histopathological examination, and pre-operative planning for proper management.Wide excision with tumor free margins may be adequate in the management of abdominal wall tumors.

5.References

children had improved cervical rotation or head tilt after the injections [19]. Futhermore, Joyce et al. reported high satisfaction scores after an average follow-up length of 22 months in the treatment of recalcitrant CMT by Botox. Their study shows that there may be a potential role for Botox in the future of CMT [20].

4.Conclusion

This particular study demonstrates that congenital muscular torticollis of the left SCM resulted from fibromatosis colli (pseudo-tumor). This case shows that controlled manual stretching and active positioning stimulation are safe and effective in the treatment of congenital muscular torticollis of the left SCM. There was an overall improvement in the infant's neck range of motion and head position.

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Conflict of Interest:

The authors declare that there is no conflict of interest

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